RELATO DE CASO

GIANT RIGHT ATRIAL MIXOMA WITH IMPORTANT HEMODYNAMIC REPERCUSSION IN CHILD: A CASE REPORT

MIXOMA ATRIAL DIREITO VOLUMOSO COM IMPORTANTE REPERCUSSÃO HEMODINÂMICA EM CRIANÇA: RELATO DE CASO

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ABSTRACT

Cardiac tumors are uncommon conditions in medical practice, and among them, the most prevalent is the myxoma. Despite their benign nature, myxomas may have behavior suggestive of malignancy and serious consequences. They may have cardiovascular repercussions associated with nonspecific systemic manifestations, which often hinder their diagnosis, benefiting their evolution and increasing the risks of serious complications, including embolic alterations and death. We present, in this work, the case report of a child admitted to the Hospital Infantil Público de Palmas (HIPP), with clinical history of abdominal distension, adynamia, hyporexia and intermittent fever, initially diagnosed with right atrial myxoma. The patient had different epidemiological characteristics than those described in the literature for cardiac myxomas. This, combined with the fact that cardiac myxomas have a low incidence and commonly present in non-specific cardiovascular and constitutional manifestations, made it difficult to diagnose and delay surgical treatment. Attention to the possibility of this disease and the echocardiogram in the evaluations may facilitate the early diagnosis of this benign cardiac neoplasia.

Keywords: Myxoma; Echocardiography; Neoplasms.

RESUMO

Os tumores cardíacos são afecções pouco comuns na prática clínica, dos quais o mais prevalente é o mixoma. Apesar de seu caráter benigno, os mixomas, podem ter comportamento sugestivo de malignidade e consequências graves. Podendo cursar com repercussão cardiovascular associada a manifestações sistêmicas inespecíficas, que muitas vezes dificultam o seu diagnóstico, beneficiando a sua evolução e elevando os riscos de graves complicações, incluindo alterações embólicas e morte. Apresentamos, nesse trabalho, o relato de caso de uma criança admitida no Hospital Infantil Público de Palmas – TO (HIPP), com história clínica de distensão abdominal, adinamia, hiporexia e febre intermitente, inicialmente diagnosticada com mixoma atrial direito. Tal paciente apresenta características epidemiológicas distintas do descrito na literatura para mixomas cardíacos, isso, associado ao fato de os mixomas cardíacos apresentarem baixa incidência e comumente evoluírem com manifestações cardiovasculares e constitucionais inespecíficas, dificultaram seu diagnóstico e retardaram o tratamento cirúrgico. A atenção com a possibilidade de ocorrência desta doença e a realização do ecocardiograma nas avaliações pode facilitar o diagnóstico precoce desta neoplasia cardíaca benigna.

Palavras-chave: Mixoma; Ecocardiografia; Neoplasias.

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INTRODUCTION

The cardiac tumors are rare diseases in clinical practice and can be classified according to their origin in primary or secondary, and these are most common in a ratio of 40:1. Among the primary ones, it is possible to classify them as benign or malignant, of which the most common is the myxoma.[1]

Histologically, myxomas are benign tumors derived from multipotent mesenchymal cells of the subendocardium.[2]These tumors are rare, with an estimated overall incidence of 0.5 cases per 1 million population per year and represents approximately 70% of all cardiac tumors.[3]

Despite their benign nature, myxomas may have behavior suggestive of malignancy and serious consequences.[1] Patients with myxoma may be asymptomatic in 46.4% of cases. However, the symptoms of this neoplasm, when present, are non-specific and vary with tumor location, size and mobility.[3] They may have cardiovascular repercussions associated with nonspecific systemic manifestations, which often hinder their diagnosis, benefiting their evolution and increasing the risk of serious complications, including embolic alterations and death. The rapid diagnosis and surgical treatment provide a favorable prognosis for patients with these tumors.[4]

We present here the case of a child admitted to the Hospital Infantil Público de Palmas (HIPP), with a history of abdominal distension, adynamia, hyporexia and intermittent fever, initially diagnosed with right atrial myxoma.

CASE REPORT

A.S., 7 years, male, brown, natural and from Formoso do Araguaia, Tocantins(TO). He was referred to the HIPP in November 2017, with abdominal distension, adynamia, hyporexia and intermittent fever for 1 month. His mother reported that the symptoms started after a day of recreational activity at school. After this event, the child developed abdominal distension and constipation, concomitant with signs of infection of the upper respiratory tract (fever, cough and hyaline coryza). After one week he improved his condition, but he developed an intermittent fever. He sought medical attention at the time, which was prescribed sulfamethoxazole/trimethoprim and simethicone, both for 7 days.The proposed treatment was performed; however, the child did not present clinical improvement, remaining with adynamia, hyporexia, abdominal distension and paleness. It was evaluated by two more general practitioners who did not reach a diagnosis. Due to the worsening of the condition, he sought pediatric medical care, where the presence of an audible heart murmur was evidenced in all points of cardiac auscultation, an echocardiogram was requested and later referred to the HIPP. Mother denies any intercurrences during pregnancy and delivery, comorbidities, previous surgeries, previous hospitalizations. Presented updated vaccination card. Family history without similar events.

In the physical examination, at the time of hospital admission, the patient was in regular general condition, hypoactive, sleepy, pale (4+/4+), eupneic, afebrile, acyanotic, anicteric and with mild face edema. He presented a normal respiratory auscultation, a "tumor plop" sound at the heart auscultation and bilateral mild jugular swelling at the cardiovascular examination. His abdomen was distended, painful to deep palpation, palpable liver at 6cm from right costal border, nonpalpable spleen, flat umbilical scar and bowel sounds were presents. His lower limbs did not have edema.

Supplementary examinations at admission: hemoglobin: 8.1 g/dL with microcytosis and hypochromia, hematocrit: 28.5%, leukocytes: 11.240/mm^3 platelets: 448,000/mm^3; C-reactive protein (CRP): 114.22mg/L; Albumin: 3.20g/dL; Globulins: 3.65g/dL. An electrocardiogram showed a sinus tachycardia, incomplete right bundle branch block and right atrial hypertrophy (Figure 1). The echocardiogram evidenced a right atrial myxoma obstructing the right ventricle entryway, with significant hemodynamic repercussion; increased right cavities; preserved left ventricular systolic function (Figure 2A and 2B). An abdominal ultrasonography showed homogeneous hepatomegaly and small accessory spleens.

During hospitalization at HIPP the patient made use of symptomatic medications, iron supplementation and was submitted to hemotransfusion, with improvement of the adynamia. Infectious screening was performed with extensive investigation, including visceral leishmaniasis, due to inversion of the albumin-globulins ratio, hepatomegaly, and intermittent fever that the patient presented in the period. However, no focus of infection were found.

After 34 days hospitalized in HIPP, the patient was referred to the Instituto de Cardiologia do Rio Grande do Sul (ICRS), in Porto Alegre, Rio Grande do Sul (RS). On the fifth day of hospitalization at the ICRS, he underwent resection of the intracardiac tumor and atrioseptoplasty. The analysis of the resected material showed a benign pericardial cyst compatible with mesothelial cyst measuring 1.5 cm in its largest diameter and cardiac myxoma with areas of hemorrhage, weighing 41.0 g and measuring 3.5 cm in its largest diameter.

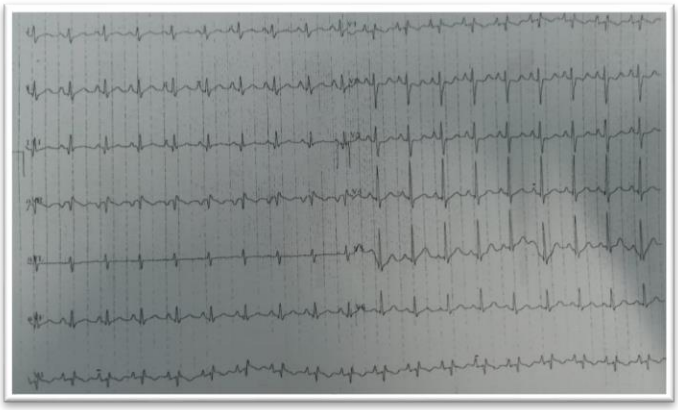

Figure 1: Electrocardiogram showing sinus tachycardia, incomplete right bundle branch block and right atrial hypertrophy.

A chest X-ray performed in the first postoperative period (PP) evidenced mediastinum without alterations and absence of pleuropulmonary lesion in activity (Figure 3). Echocardiogram performed in the third PP showed moderate tricuspid insufficiency, mild mitral insufficiency and right cavity enlargement.

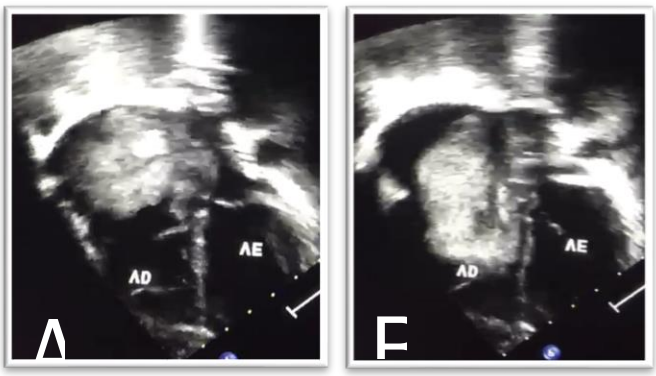

Figure 2A: Echocardiogram demonstrating a bulky mass almost completely filling the right atrium during ventricular systole. **Figure 2B:** Echocardiogram demonstrating tumor mass protrusion by the right atrioventricular valve during ventricular diastole.

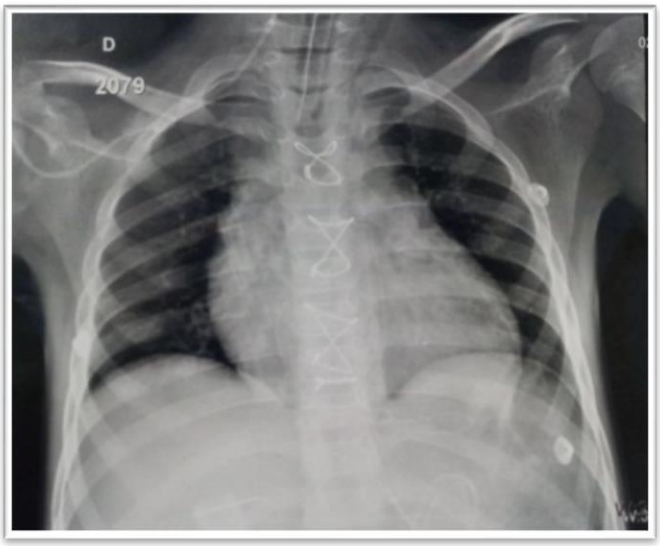

Figure 3: Postoperative chest X-ray evidencing mediastinum without alterations and absence of active pleuropulmonary lesion.

The patient was discharged in the 15th PP with good clinical and surgical evolution. After discharge, he returned to the city of origin and maintained an outpatient follow-up.

After six months of the surgical procedure, the patient returned to the outpatient cardiopediatric clinic without symptoms, with systolic murmur in tricuspid focus (1+/6+) on cardiac auscultation. Abdominal physical examination showed no palpable liver below the costal border. The echocardiogram performed at the day showed mild tricuspid insufficiency, slight right atrial enlargement and good biventricular function.

DISCUSSION

Cardiac myxomas are the most common primary benign neoplasms of the heart.[5] The most common site of these tumors is the left atrium in about 75% of cases, followed by the right atrium and ventricles, rarely appearing in the heart valves.[1,5-8] These tumors have a higher incidence in women between the third and fourth decade of life and are rare in children.[1,4]

They are usually pedunculated, sporadic and solitary tumors, but when associated with rare familial forms of cardiac myxomas, they preferentially affect younger individuals and may present in a multicentric form in up to half of the cases.[1,7-8]

The growth of tumor mass in sporadic forms may range from 1.3 to 6.9 mm/month, measuring 5 cm on average, and their average weight is 37g, reaching up to 180g.[8]

This patient's case can be considered rare and therefore becomes relevant because it contradicts several epidemiological parameters. The patient was a male child with tumor located in the right atrium, but the characteristics of the mass were within the expected: pedunculated, solitary and weighed 41g. Another detail to highlight is the patient's young age. This data might suggest a congenital characteristic, but a careful questioning about the family history did not reveal any similar case in their relatives.

Patients with myxoma may be asymptomatic in almost 50% of the cases.[3]However, although the tumor has benign characteristics the symptomatology will depend on the size, shape, mobility and location of the tumor.[1,3,4,8] When present, their manifestations are diverse and nonspecific, making a differential diagnosis with other heart diseases or systemic diseases.[1,3]

Symptomatic cardiac myxoma patients present with at least one of the characteristics of a classically described triad, which includes: the hemodynamic consequences of intracardiac obstruction, embolic events, and nonspecific constitutional or systemic symptoms.[3-5,8]The most common manifestations are dyspnea, atypical chest pain and syncope.[8] They may also present cardiac arrhythmias due both to direct infiltration of cardiac conduction tissue or to myocardial irritation.[5]

Hemodynamic changes are the most common. They may be intermittent and manifest in certain patient positions in which the tumor mass projects into the mitral or tricuspid valves going into the ventricles or in more advanced cases have more chronic consequences. Symptoms may range from episodes of dyspnea, arrhythmias (mainly atrial fibrillation), palpitations, chest discomfort, dizziness, syncope, heart failure, acute pulmonary edema and sudden death.[3,4]In younger and male patients, neurological symptoms tend to be more evident, while in female patients the systemic symptoms tend to be more expressive.[3]

The right atrial myxoma can obstruct the tricuspid valve, causing symptoms of right heart failure, peripheral edema, hepatic congestion and syncope, as evidenced in the case reported.[5]

Embolic events are very serious or even fatal complications of myxomas and present a higher incidence when the tumor is in the left atrium.[4] The main embolic complications reported are ischemic stroke, transient ischemic attack, acute myocardial infarction, acute pulmonary embolism and peripheral embolisms.[3] These embolic events may result from the formation of clots on the tumor mass or the detachment of fragments from the tumor.[9,10] In the case reported, the child did not present embolic phenomena.

Constitutional symptoms are systemic manifestations, which often resemble autoimmune disease such as fever, weight loss, arthralgia, asthenia, adynamia, myalgia, and

fatigue.[3,8,11] These symptoms occur through the production of Interleukin-6 by the tumor. The clinical and immunological normalization in these patients is accompanied by regression in the levels of this interleukin.[8,11]

Cardiac auscultation in atrial myxomas may vary with his size, location, mobility, prolapse through the atrioventricular valves and even with body position, so that cardiac murmur detection may not occur.[4] However, among the signs, the most significant and characteristic of myxoma to cardiac auscultation is the so-called "tumor plop" heartsound - noise that the tumor makes when passing through the atrioventricular valves.[1,3,4]

The patient in the reported case had an important cardiac murmur ("tumor plop") which was audible in all points of cardiac auscultation and corresponded to a murmur of 4+/6+ (high murmur with tremor). In addition, he manifested constitutional symptoms and hemodynamic changes. The myxoma present in the right atrium behaves as a right heart failure resulting in congestion and difficulty in the systemic venous return, which can result in portal hypertension and, consequently, hepatomegaly, perceived in the patient.

Because of its wide variety of symptoms, cardiac myxomas can make a differential diagnosis with cardiac valvular disease, cardiac insufficiency, intrinsic pneumopathy, pulmonary arterial hypertension, cardiomegaly, cerebrovascular disease, rheumatic fever, bacterial endocarditis, myocarditis, vasculitis, supraventricular and ventricular rhythm disorders , syncope, pulmonary and systemic embolisms and other diseases.[4, 8]

The echocardiogram has a high positive predictive value in the diagnosis of cardiac myxomas, being the gold standard for the diagnosis. It provides information on the size, location, fixation, mobility and prolapse through the atrioventricular valve, in addition to allowing the study of other cardiac cavities due to the possibility that these tumors are not solitary, making it useful also in evaluating tumor recurrence.[1,4,8]Other imaging exams such as cardiac catheterization, computed tomography and magnetic resonance imaging detect these tumors and may provide additional information as possible associated complications.[4]

The chest X-ray and electrocardiogram are non-specific, however this is critical to assess arrhythmias or cardiac blockages by direct infiltration of the tumor into the cardiac conduction tissue or own myocardial irritation.[4,8] The electrocardiogram of the patient in this clinical case demonstrated sinus tachycardia and incomplete right bundle branch block, changes that, although relevant, are nonspecific.

Laboratory tests also present nonspecific findings in cardiac myxomas, such as: elevation of erythrocyte sedimentation rate, thrombocytopenia, polycythemia, anemia, elevated globulin levels, increased CRP and leukocytosis.[1,4] As evidenced in the clinical case reported, the presence of significant anemia, mild leukocytosis, reversal of albumin-globulin pattern and persistent increase in CRP.

As soon as the myxoma diagnosis is made, the surgical management should be scheduled immediately because of the high risk of thromboembolic events and possible fatal outcomes.[3,8] The treatment of choice is complete surgical resection of the tumor, which in most cases is curative.[1,3,12] Regardless of the technique used to perform resection, the tumor should be minimally manipulated during the procedure to avoid embolic events.[3]

As surgical treatment is usually definitive, and the rate of recurrence is very low (about 3% in sporadic tumors), the presence of recurrence should alert to the presence of familial myxomas, incomplete resection and transition to malignancy, events commonly associated with recurrent tumors.[8,13] After initial resection, recurrence may occur within a few months to several years.[3,13]

Therefore, regular long-term follow-up is recommended in all patients with cardiac myxoma, particularly in patients with a familial form of cardiac myxoma. Follow-up should involve medical history, physical examination and periodic echocardiographic studies.[3,8,11] The patient in this report performs regular half-yearly follow-up with echocardiography and consultations in an outpatient cardiopediatric clinic.

CONCLUSION

The fact that cardiac myxomas present a low incidence and commonly evolve with non-specific cardiovascular and constitutional manifestations makes diagnosis difficult and delays surgical treatment. The attention to the possibility of occurrence of this disease and the completion of the echocardiogram in assessments can facilitate the diagnosis of this cardiac benign neoplasia, and, early diagnosis and surgical treatment improve the prognosis of patients, avoiding serious and even fatal complications.

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